Case Report

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An accidental finding of a retroperitoneal desmoid tumor: case report and review of the literature

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Abstract

Retroperitoneal desmoid-type fibromatosis is a rare benign mesenchymal neoplasm that develops as a result of fibroblastic proliferation within the musculoaponeurotic stroma. The authors present the case of a 41-year-old male patient who was referred for a retroperitoneal neoplasm. A mesenteric mass core biopsy was done, and it revealed a low-grade spindle cell lesion consistent with desmoid fibromatosis.

Keywords: desmoid, intra-abdominal mass, mesenteric mass

Introduction

Retroperitoneal desmoid-type fibromatosis (DTF) is a rare benign mesenchymal neoplasm that develops as a result of fibroblastic proliferation within the musculoaponeurotic stroma[1,2]. This tumor may infiltrate the surrounding tissue, but hardily metastasize distantly[3]. DTF can be extra-abdominal or intra-abdominal, with the former being more common[4]. It is characterized by a locally aggressive infiltrative growth manifesting on cross-sectional imaging as a well-circumscribed or poorly defined solid mass[1]. DTF is found to occur in multiple locations, but they most commonly arise from the abdominal wall, mesentery, and the extremities followed by the abdominal cavity and retroperitoneum[4,5]. It can be hereditary or sporadic, with the latter being associated with familial adenomatous polyposis (FAP) or Gardner's syndrome[2]. Incidence has been reported to occur in 7.5–16% of FAP patients or those with Gardner's syndrome. These patients have a higher relative risk of a developing desmoid tumor than the general population, which necessitates screening. Tumors associated with FAP are more likely to develop in the intra-abdominal area[5]. Desmoid tumors most commonly affect young adults between the ages of 25–35. Studies have shown a female to male predilection of a 2 : 1 ratio[2,4]. Rarely, desmoid tumors may demonstrate aggressive proliferative activity, which can lead to malignant complications secondary to the mass effect. As a consequence, they can cause bowel ischemia or intestinal obstruction[2]. The exact etiology of fibromatosis is not yet fully known; however, it is usually associated with endocrine factors or trauma[3]. We herein report a case of an incidentally discovered retroperitoneal desmoid tumor, which was preoperatively considered to be an intra-abdominal desmoid tumor. To reach an accurate diagnosis and an effective treatment plan, we must elucidate this atypical presentation.

HIGHLIGHTS

- Retroperitoneal desmoid-type fibromatosis is a rare benign mesenchymal neoplasm that develops as a result of fibroblastic proliferation within the musculoaponeurotic stroma.
- We present the case of a 41-year-old male patient who was referred for a retroperitoneal neoplasm.
- A mesenteric mass core biopsy was done, and it revealed a low-grade spindle cell lesion consistent with desmoid fibromatosis.

Literature review

Case presentation

This is the case of a 41-year-old male patient who was referred for a retroperitoneal neoplasm. The retroperitoneal mass was accidentally discovered by an abdominal MRI scan. CA19-9 and CA125 were done, which returned normal. A mesenteric mass core biopsy was done, and it revealed a low-grade spindle cell lesion consistent with desmoid fibromatosis (DF). The physical

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examination was unremarkable, and the patient was vitally stable. The blood type is O + and is negative for antibodies. As an initial diagnosis, an intra-abdominal desmoid tumor was suspected. The main line of management was proposed to be a laparoscopic resection of an intra-abdominal desmoid tumor with laparoscopic resection anastomoses of the small bowel. A series of lab tests were done starting with a complete blood count, which showed that the erythrocyte sedimentation rate was within normal range, white blood cell levels were low (3.71×10). Whereas, red blood cells are within normal range, and hemoglobin level is normal. Furthermore, hematocrit, mean corpuscular volume, mean corpuscular hemoglobin, mean corpuscular hematocrit, red cell distribution width, and mean plasma volume are all within normal ranges. Platelets were found to be low (144 k/μl). Segment neutrophils are normal; however, absolute neutrophils are low (1.67 k/μl). Lymphocytes are high (47.30%), while absolute lymphocytes are normal. Monocytes and absolute monocytes are normal. Eosinophil count and absolute eosinophils are normal. While basophil counts are low (0.19%), absolute basophil is normal. In the basic metabolic test, serum urea was elevated (38.52 mg/dl) and serum creatinine (1.53 mg/dl) was slightly elevated; the remainder of the results were normal. A coagulation profile was done and showed elevated partial thromboplastin time (36.50 s) and prothrombin time in citrated plasma (13.70 s), while the international normalized ratio was normal. Serology tests were found to be unreactive. The abnormal findings can be found summarized in Tables 1 and 2.

For histopathological analysis, a specimen was obtained via a computed tomography (CT)-guided biopsy and paraffin-embedded block. Gross picture showed two tiny soft tissue cores measuring 0.5 cm each, embedded all in one cassette. A mesenteric mass, core biopsy, and low-grade spindle cell neoplasm consistent with DF were the results. Immunohistochemistry showed that target cells are positive for β-catenin (nuclear staining) and SMA, while negative for S100, desmin, CD34, EMA, CD117, and ALK-1.

As an initial test an erect abdominal radiography was done which revealed multiple centrally dilated small bowel loops with air fluid level and large colon collapse, a left hypochondrial air strip suggestive of pneumoperitoneum was also noted. All this gave an impression of small bowel obstruction with suspected pneumoperitoneum for CT correlation. An abdominal/pelvic ultrasound was done to obtain more details, which showed that some of the bowel loops in the lower abdomen are dilated up to 3.4 cm showing back and forth movements with minimal free fluid in between. It also showed a few intraluminal stones in the gallbladder, with the largest being 10 mm; hence, chronic calcular gallbladder disease was also suspected. An abdominal multidetector CT (MDCT) with and without contrast was done since the ultrasound returned with positive findings. MDCT without contrast showed a mild line infraumbilical abdominal mass lesion is seen showing internal heterogeneity, measuring 7.4×4.2 cm. It is seen displacing the small bowel loops with encasement of one of the branches of the superior mesenteric artery; a preserved clear fat plane with the sigmoid colon is noted. Minute upper pole renal cysts bilaterally are noted too. This was followed by an MDCT with contrast which showed small basal atelectasis in the lower chest. Two uncomplicated gallbladder stones and in the bowel diffuse mild small bowel dilatation seen (3 cm) with air fluid level with small bowel feces seen. The oral contrast is seen reaching the distal jejunal and proximal ileal loops. A small bowel anastomosis is seen in the right iliac fossa. No definite transition seen. Postoperative changes along the small bowel mesentery. These findings can be seen in Figure 1a and b.

Another CT without contrast was done later to compare with the previous one. The recent CT revealed an increase in the degree of small bowel dilatation compared to the previous study, with a transitional zone around the anastomotic site between a dilated small bowel loop and a relatively collapsed terminal ileum. This would collectively be in favor of partial distal small bowel obstruction around the site of anastomosis at the distal ileum at the right iliac fossa, warranting clinical and lab correlation and follow-up assessment. Less likely radiological differences include abnormally prolonged postoperative ileus related to recent small bowel anastomosis. There are no signs of related complications. Otherwise, the studies are stable compared to the last CT scan, with no newly developed abnormalities or significant interval changes. To be more accurate, an MRI was also performed, the results of which can be seen in Figure 1C–E. After all these tests, a diagnosis of an intra-abdominal desmoid tumor was established. The plan of management was to do a laparoscopic resection of the intra-abdominal desmoid tumor with laparoscopic resection anastomoses of the small bowel. Postoperatively the patient was monitored closely and then discharged after 3 days of stay.

Discussion

A desmoid tumor, as defined by the WHO in 2002, is a desmoid-type deep fibromatosis[11], where a 'fibromatosis' consists of proliferation of well-differentiated myofibroblasts in a collagenous extracellular matrix. They are benign, locally aggressive (thus nonmetastatic), mesenchymal tumors composed of fibrous elements. They can be classified anatomically as mesenteric or pelvic, intra-abdominal, extra-abdominal, or tumors within the abdominal wall[12]. Extra-abdominally, they usually manifest in the shoulder, chest wall, back, thigh, head, neck, pelvic girdle, or glutes[13,14]. Retroperitoneal intra-abdominal DF make up for less than 1% of retroperitoneal masses, yet 28–69% of desmoids generally[15] Abdominal DF can be further subclassified as superficial (fascial) or deep (musculoaponeurotic) based on their connective tissue origin[13,16].

Epidemiologically, DF is rare, yet the most common primary mesenteric tumor[17]. It carries an incidence of 3.7 new cases per million people per year, and accounts for 0.03% of all neoplasms and less than 3% of all soft tissue tumors[15]. It is prevalent in 13% of patients with FAP coli and in 4–29% of Gardner syndrome patients[14,15]. Age-wise, they may occur in all age groups but are typically seen between 30 and 50 years of age[15]. Furthermore, any gendered, racial, or ethnic distinction is insignificant, albeit it slightly favors females[17]. The causal mechanisms for desmoids are debatable and yet to be determined, albeit it is multifactorial due to the presence of several commonplace risk factors, including hormonal factors, especially hyperesterogenic states such as estrogen therapy or oral contraceptive use[12,16], pregnancy, and genetic abnormalities, thus potentially explaining the slightly higher predilection to females of reproductive age. However, a relatively recent review disputed such a finding by revealing that postoperative recurrence was not associated with sex, nor was pregnancy associated with an increased risk of tumor progression or recurrence after surgery[18]. Trauma and previous surgery also account on average for 19–63% of desmoid cases, 50% of which developed in the first 4 years after the

Table 1

Past studies reporting cases of retroperitoneal fibromatosis desmoid tumor

References	Age and sex	Chief complaint	Location of lesion	Radiologic modality for diagnosis confirmation	Radiological findings	Treatment
Lee *et al.*[1]	46, Male	Left lower quadrant pain and a palpable mass in the left upper abdomen	Left upper abdomen	Plain radiographs Ultrasonography contrast-enhanced (CT) scan	Large mass-like opacity in the left abdomen Large, thick-walled cystic mass without evidence of an intracystic solid portion or septum 18.3 × 12.3 × 21.5 cm sized oval cystic mass with a relatively thick wall in the left upper abdomen	Laparoscopic spleen-preserving distal pancreatectomy without preoperative biopsy due to a risk of rupture
Xiao *et al.*[3]	28, Male	Tangible abdominal mass in the right middle abdomen	Lower abdomen, under the ileocecal mesentery	CT scan	Mass with uniform soft tissue density and inhomogeneous enhancement in lower abdomen; the maximum cross-sectional area of the mass was about 7.1 × 7.6 cm	Tumor mass resection, right colon resection, partial duodenum resection, and intestinal anastomosis
Zhang *et al.*[6]	60, Male	Abdominal mass for 6 months	Left side of the old surgical incision for gastric cancer	Ultrasound Enhanced CT scan	Mass of 67 × 13 × 25 mm with unclear boundary Mass involvement of the left rectus abdominis, and malignancy could not be ruled out	Surgical resection along 5 cm of the surrounding mass
Campara *et al.*[7]	35, Male	Frequent urination with the feeling of pressure and pain	Front wall of the bladder with diameter of 70 mm with signs of infiltration of the musculature of the anterior abdominal wall	Ultrasound CT scan	Tumor change the largest diameter to 70 mm, which covers the front wall of the bladder, and whose origin cannot be determined Edge well vascularized formation that covers the front wall of the bladder, with visible signs of infiltration of the musculature of the anterior abdominal wall measuring 75 × 80 mm	Resection of pelvic tumors with partial cystectomy and right-hand iliac lymphadenectomy
Shih *et al.*[8]	56, Male	Epigastralgia for 3 weeks	Retroperitoneum adhering to the peritumor vessels, nerves and the pancreatic tail	CT scan	Progressive enlargement of the splenic hilar tumor from 1.9 to 3.2 cm	Resection en bloc with sacrifice of adjacent vessels and nerves
Aidid *et al.*[9]	31, Female	Recurring abdominal pain	Right retroperitoneal cavity	CT scan	Well-circumscribed mass demonstrating soft tissue density and homogeneous enhancement after contrast, associated with fat stranding all around	Total colectomy with ileoanal anastomosis and removal of the retroperitoneal mass infiltrating the inferior duodenal flexure
Ghidirim *et al.*[10]	54, Male	Palpable abdominal mass	Left abdominal retroperitoneum	Abdominal ultrasonography, CT scan	Solid mass with relatively well-defined borders	Complete mass resection

Table 2

Abnormal lab results

Variables	Normal values	Results
Hematological parameters		
White blood cells (k/μl)	4.1–10.8	3.71
Platelets (k/μl)	150–400	144
PTT (s)	25–35	36.50
Prothrombin time (s)	11–13.5	13.70
Differential		
Absolute neutrophil (k/μl)	2.5–6.5	1.6
Lymphocyte (%)	24–44	47.3
Renal parameters		
Serum creatinine (mg/dl)	0.74–1.35	1.53
Serum urea (mg/dl)	5–20	38.52

PTT, partial thromboplastin time.

predisposing incidents[19,20]. Genetic anomalies and syndromes are also major risk factors, especially in regards to genetic mutations of the adenomatous polyposis coli gene such as in FAP or Gardner's syndrome, in the beta-catenin gene (CTNNB1), or other mutations in molecular interlinks between the wound healing process and mesenchymal fibroproliferative tumor development[12,19]. However, the pathogenesis of DF is not completely understood overall.

Histologically, DF is usually formed of uniform and elongated spindle-shaped or stellate fibroblastic cells in the background of collagenous stroma[17,21], in the absence of a pseudocapsule, which increases the tendency to infiltrate[18]. Generally, this is present in absence of any neuronal or muscular differentiation with little to no evidence of inflammation[21].

On gross examination, desmoid tumors are circumscribed lesions with irregular or infiltrative borders[22]. They usually present as a large lesion measuring from 5 cm to even greater than 15 cm. DF can present as multiple tumors in 10–15% of cases[21]. Cut surface of a desmoid tumor might take a white or tan color with fibrous trabecular resembling to that of a scar tissue[21,22].

Patients with DF usually presents with symptoms based on the site where the tumor arises from, these sites can be divided into: abdominal wall, intra-abdominal, and extra-abdominal[23]. In cases of intra-abdominal desmoid tumors, usually the patient remains symptom-free until the tumor growth or nearby invasion causes vascular, ureteric, intestinal, or neuronal symptoms[20]. Pain is the most common presenting complaint and can be abdominal, pelvic, leg, or vulval[24]. Other signs can include weight loss, fever, nausea, and vomiting[14,17,23]. Desmoid tumors may be clinically misdiagnosed as ovarian, mesenteric, or other retroperitoneal tumors but the lack of pleomorphism, cellularity, and mitotic activity makes these tumors easily distinguishable from sarcomas on the basis of histological inspection[25]. Desmoid tumors have local invasive properties and usually tend to affect blood vessels. Despite having locally aggressive fibroblastic growth properties, DF tumors lack metastasizing characteristics and are histologically classified as benign tumors[9]. Goulding and colleagues reported on a case of DF resection where there was an intraoperative discovery of invasion of the external iliac vein and internal iliac vessels. In order to remove the tumor, the researchers sacrificed the vessels.

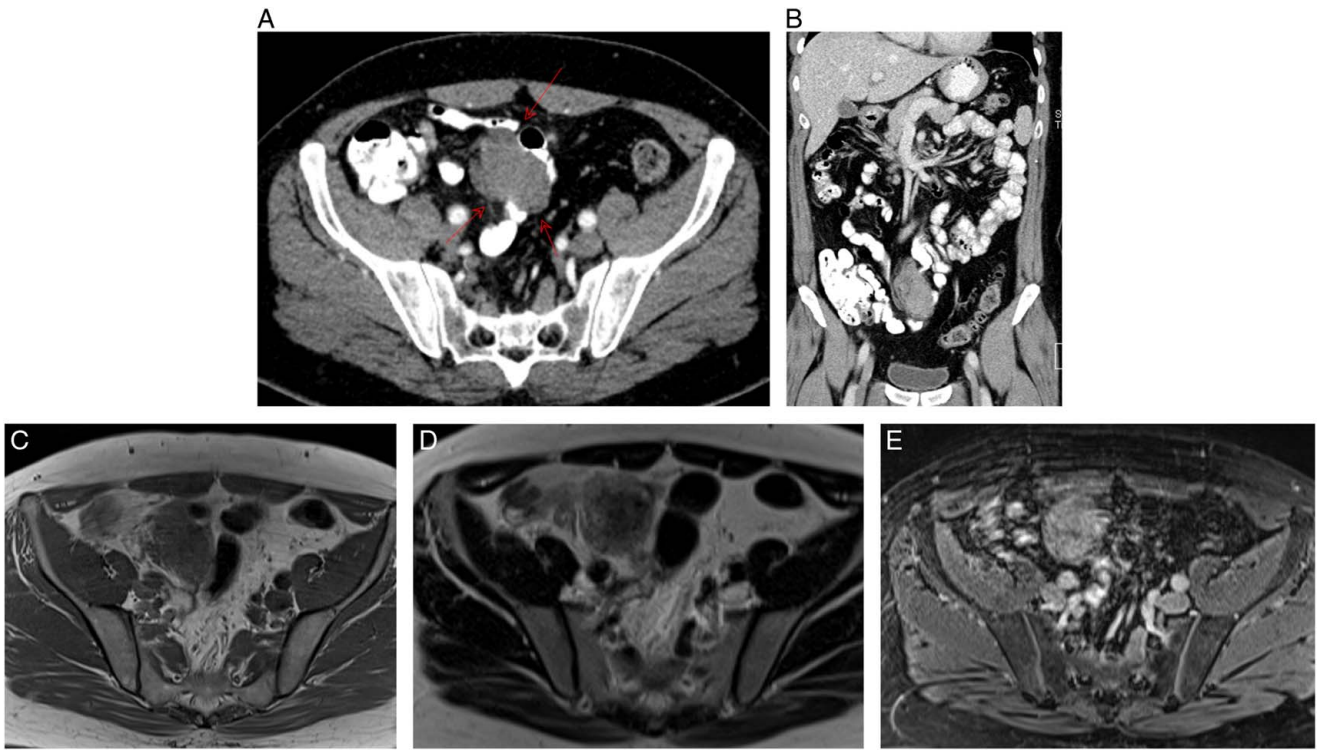

Figure 1. Postcontrast computed tomography axial (a) and coronal (b) images show a mesenteric soft tissue attenuation mass lesion with lobulated outlines displacing the bowel loops measuring 5.5 × 3.5 × 4.5 cm in maximum dimensions. MRI axial T1 (c), T2 (d), and postcontrast VIBE (e) show intermediate signal intensity of the lesion in T1 and T2 sequences with mild postcontrast enhancement. VIBE, volumetric interpolated breath-hold examination.

Consequently, we must prepare for the prospect of sacrificing any nearby vessels or surrounding structures[26].

In terms of investigations, an abdominal or pelvic ultrasound was done and showed that some of the bowel loops in the lower abdomen are dilated up to 3.4 cm, showing back and forth movements with minimal free fluid in between. It also showed a few intraluminal stones in the gallbladder, with the largest being 10 mm; hence, chronic calcular gallbladder disease was also suspected. An abdominal MDCT with and without contrast was done afterwards, clearly showing borders and tissue density compared to plain scanning methods, and was then followed by an MRI, as it proved valuable in determining the size, composition, and infiltration of the surrounding tissue, as well as identifying the tumor envelope or edema around the tumor. MRI is the preferred imaging methods for evaluating the tumor preoperatively and detecting recurrence postoperatively[27].

As for treatment options, they include pharmacological agents such as c-AMP modulators (theophylline, chlorothiazide, ascorbic acid, and testolactone), which did not show convincing results due to the low mitotic activity seen in the tumor, hindering their efficacy[28,29]. Tamoxifen, however, showed better results. It has an antiestrogen action that inhibits fibroblast growth in the tumor[30,31]. Tamoxifen is also preferred over steroids and cytotoxic drugs because it causes fewer side effects and is the best choice in young women as it decreases the rate of hemorrhage and improves the chances of fertility. Other treatment options include surgery with or without radiotherapy or chemotherapy[32], the latter of which has proven to show clinical benefit[33]. In contrast, the efficacy of radiotherapy after surgery or for unresectable tumors is yet to be proven[34]. Most researchers conclude that the best option for treatment of desmoid tumor is total excision of the primary tumor, although resection with positive margins may have a high recurrence rate in the future, which can reach up to 40%. Re-resecting following a second or third trial of excision will not change the recurrence rate, and it may even be equal to the first attempt[35]. No recurrence was found after surgical removal of the tumor with eight cases, which have been followed up for 2 years[36].

Differential diagnoses of DF are often considered first due to the scarcity of desmoids, which are considered among the last differentials diagnoses of a pelvic or abdominal mass. Nonetheless, they include solitary benign fibroblastic proliferation and reactive processes or low-grade myofibroblastic sarcomas, both of which can also be identified by nuclear β-catenin staining, gastrointestinal stromal tumors (by being c-kit positive), or even desmoids of other locations. Clinically, they can also be mistaken for ovarian, mesenteric, or retroperitoneal tumors[26,37].

Due to the rarity of the condition, it is difficult to study its natural history, and thus prognosis is difficult to assign, added to the fact of the prognostic variability of desmoids, as some tumors may spontaneously regress, others may be slow and indolent, and others may be progressive and aggressive[26]. Moreover, their locally invasive nature can create operative difficulty as well as the necessity of sacrificing certain important adjacent structures such as the iliac vessels, ureter, or even pancreas; such findings may not be identified by imaging preoperatively, thus increasing the need for prompt intraoperative decision making and excision on the spot, which is no doubt a challenging ordeal[26,38]. The invasive nature of desmoids can also cause a myriad of complications, whether speculative or defined in literature, such as small bowel obstruction, hydronephrosis, ureteric obstruction or rupture, intestinal perforation, enterocutaneous fistula, and intestinal hemorrhage, each of which carries its fair share of resultant medical burdens[39]. Alternatively, morbidity and mortality with desmoids may be related to the treatments themselves, including side effects or complications of chemotherapy, irradiation, or surgery. As such, this may indicate conservative observation for some desmoids instead for a better outcome and quality of life for the patient[18,26]. To overcome such issues, early diagnosis and treatment are crucial, but this is difficult as most patients are asymptomatic[37]. Concerning regression, it can occur secondary to abolishing risk factors or presumed causative factors, such as withdrawal of estrogenic stimulation, or from secondary infarction by superimposed infection or compromised vascular supply locally[1]. In general, there is a need for more studies to be done for such cases in order to alleviate the current paucity of data regarding the behavior of desmoids to help create better guides for clinical practice and research and for better evaluation of the impact of DF treatments on quality of life formally, such as with Bertani *et al.*'s[40] paper, which used the European Organization for the Research and Treatment of Cancer (EORTC) QLQ-C30 questionnaire, yet information about early and late morbidity of surgery such as herniation, mesh pain or bulging, reoperation, short gut syndrome, and obstetric issues is still lacking[18,38].

Conclusion

In summary, we report the case of 41-year-old male patient who was referred for a retroperitoneal neoplasm. A mesenteric mass core biopsy was done, and it revealed a low-grade spindle cell lesion consistent with DF.

There is a need for more studies to be done for such cases in order to alleviate the current paucity of data regarding the behavior of desmoids and help create better guides for clinical practice and research.

Ethical approval

IRB approval was obtained from the ethical and scientific community of Batterjee Medical College for Sciences and Technology.

Consent

Participation was voluntary for all participants, and all participants had the right to withdraw at any time.

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Author contribution

All authors contributed to the present report.

Conflicts of interest disclosure

The authors declare that they have no financial conflict of interest with regard to the content of this report.

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